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Case Report

Grade III solitary fibrous tumor/hemangiopericytoma: An enthralling intracranial tumor—A case report and literature review☆

Abdul Rahman Al Armashi, MD[a,b,*], *Akram Alkrekshi, MD*[c], *Anas Al Zubaidi, MD*[d], *Francisco J. Somoza-Cano, MD*[b], *Faris Hammad, MD*[b], *Dina Elantably, MD*[c], *Kanchi Patell, MD*[b], *Keyvan Ravakhah, MD*[b]

[a] *Seidman Cancer Center, University Hospitals - Case Western Reserve University, Cleveland, OH, USA*
[b] *St. Vincent Charity Medical Center, Cleveland, OH, USA*
[c] *Metrohealth Medical Center-Case Western Reserve University, Cleveland, OH, USA*
[d] *Johns Hopkins University, Baltimore, MD, USA*

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ABSTRACT

Hemangiopericytomas account for less than 1% of all intracranial tumors. In 2016, World Health Organization (WHO) unified the two terms into a single medical condition known as solitary fibrous tumor/hemangiopericytoma (SFT/HPC). Our patient is an 80-year-old woman with a past medical history of sick sinus syndrome status post pacemaker placement. She presented to the emergency department with progressive headaches for one month duration. Her headaches worsened at night, waking her up from sleep. They also increased in intensity by bending forward. Review of systems was significant for bilateral lower extremity weakness accompanied by difficulty walking. The motor exam was remarkable for right upper and right lower extremity 3/5 weakness. The gait was ataxic. A Computed tomography scan of the head without contrast revealed a large dural-based right parietal hyperdense mass with surrounding edema, mass effect, and compression of the right lateral ventricle atrium. A right-to-left midline shift was also noted. Given the fact that our patient had a pacemaker, she was not a candidate for a brain MRI. Neurosurgery successfully resected the mass. Histopathological studies confirmed WHO grade III anaplastic solitary fibrous tumor/hemangiopericytoma. The patient was discharged on adjuvant radiation with imaging surveillance given the grade and the extent of resection. This case highlights a rare type of intracranial mass that resembles meningioma on imaging studies. It also illustrates that

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* Corresponding author.
E-mail address: Abd.armashi@gmail.com (A.R. Al Armashi).

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solitary fibrous tumor/hemangiopericytoma should be kept as a differential diagnosis for brain masses, given its aggressive nature, and its potential of metastasis and recurrence.

Introduction

Hemangiopericytomas account for less than 1% of all intracranial tumors. Previously classified as capillary angioblastic meningiomas, these tumors possess similar genetic alterations to other solitary fibrous tumors of the dura involving the neuraxis [1,2]. Consequently, in 2016 the World Health Organization (WHO) unified the 2 terms into a single medical condition known as solitary fibrous tumor/hemangiopericytoma (SFT/HPC) [3]. We present a case of a SFT/HPC presenting in an elderly female with symptoms of intracranial mass effect.

Case presentation

Our patient is an 80-year-old woman with a past medical history of nicotine use disorder and sick sinus syndrome status post pacemaker placement. She presented to the emergency department with progressive, constant, tension-like headaches that had started one month prior. Her headaches worsened at night, waking her up from sleep. They also increased in intensity by bending forward. The patient did not report any alleviating factors or previous similar episodes. Review of systems was significant for bilateral lower extremity weakness accompanied by difficulty walking, eventually unable to stand due to fear of falling. Otherwise, the review of the systems was negative for seizures, sensory loss, aphasia, blurry vision, nausea, or vomiting. Physical examination showed stable vital signs. The motor exam was remarkable for right upper and right lower extremity 3/5 weakness. The gait was ataxic. Moreover, she had unremarkable cranial nerves and sensory exam. A computed tomography (CT) scan of the head without contrast revealed a large dural-based right parietal hyperdense mass with surrounding edema, mass effect, and compression of the right lateral ventricle's atrium. A right-to-left midline shift was also noted in Figs. 1-3. Given the fact that our patient had a pacemaker, she was not a candidate for a brain MRI. Intravenous dexamethasone was promptly given while neurosurgery successfully resected the mass. Histopathological studies revealed ten mitoses per ten high-power fields on microscopy, confirming WHO grade III anaplastic SFT/HPC. The patient was discharged on adjuvant radiation therapy with imaging surveillance given the grade and extent of resection.

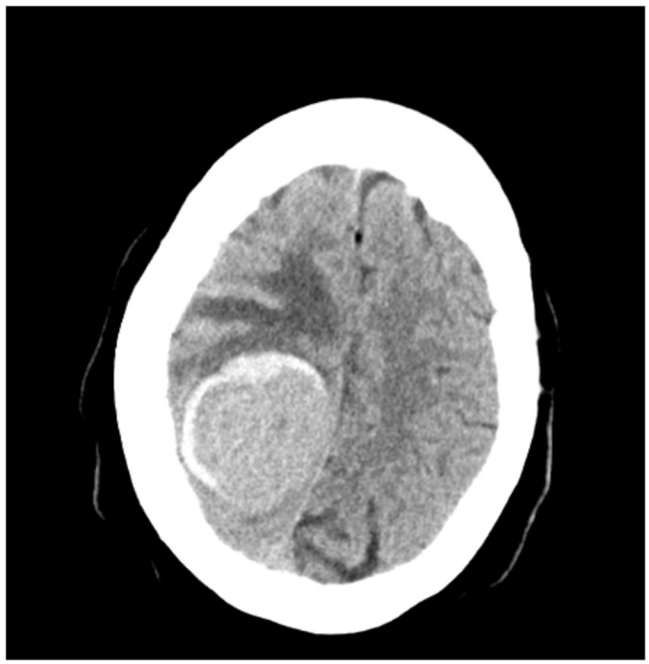

Fig. 1 – Computed tomography (CT) scan of the brain in axial view revealing a dural-based right parietal hyperdense mass with surrounding edema, mass effect, compression of the right lateral ventricle's atrium and a right-to-left midline shift.

Table 1 – WHO 2016 solitary fibrous tumor/hemangiopericytoma (SFT/HPC) grades.

WHO grade (2016) [3]	Description	Phenotype	Behavior	Prior name
I	Spindle cell lesion with high collagen content and high cellularity.	SFT	Benign tumor	SFT
II	Less collagen content, more cellularity with "staghorn" vasculature.	SFT	Malignant tumor	HPC
III	Five or more mitoses per 10 high-power fields.	HPC	Malignant tumor	Anaplastic HPC

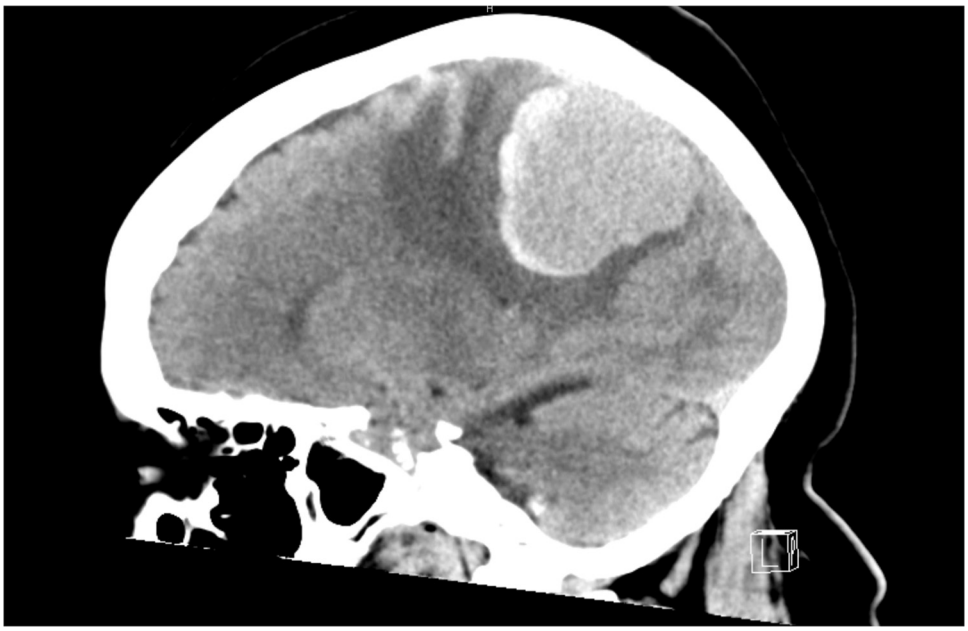

Fig. 2 – Computed tomography (CT) scan of the brain in sagittal view revealing a dural-based right parietal hyperdense mass with surrounding edema, mass effect and compression of the right lateral ventricle's atrium.

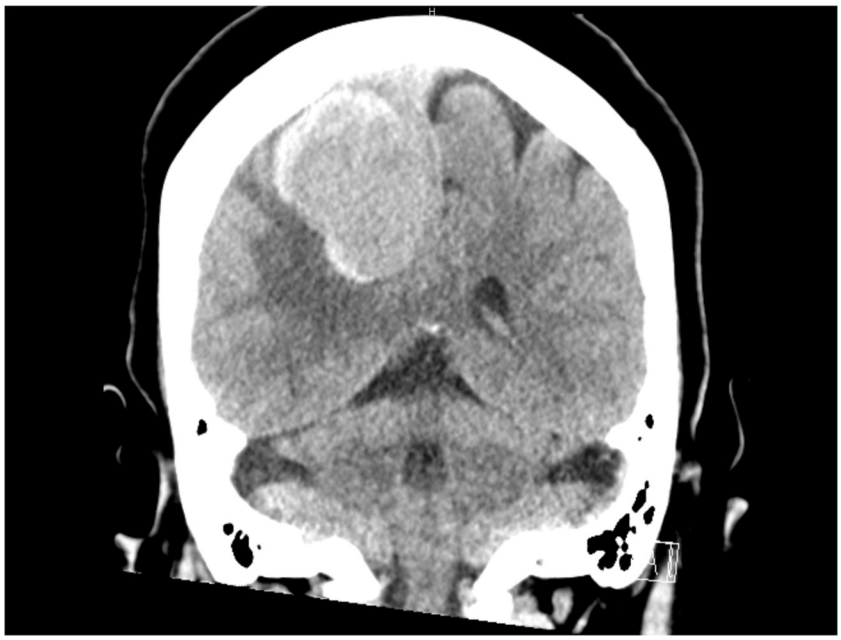

Fig. 3 – Computed tomography (CT) scan of the brain in coronal view revealing a dural-based right parietal hyperdense mass with surrounding edema, compression of the right lateral ventricle's atrium and a right-to-left midline shift.

Discussion

Solitary fibrous tumors (SFTs) are a heterogeneous category of uncommon spindle cell neoplasms that typically arise from the pleura and peritoneum. They account for less than 2% of all soft tissue masses [4]. Hemangiopericytomas are highly cellular and mitotically active tumor that is abundant in pericellular reticulin. Unfortunately, these characteristics are associated with high recurrence and metastasis [5]. Carneiro et al. described the first case of SFT/HPC discovered in the meninges in 1996 and established histological characteristics

Table 2 – Literature review of the similarities and differences between FT/HPC and meningioma based on histological and radiological findings.

Features	SFT/HPC	Meningioma
Histopathology [9,10]	STAT6 Sensitivity (96%) Specificity (100%) ALDH1 and CD34 Sensitivity (84%) Specificity (97%)	SSTR2A Sensitivity (95.2%) Specificity (92%) EMA Sensitivity (89.3%) Specificity (85%)
Imaging characteristics [11–13]	T1W1: Isointensity T2W1: Isointensity Enhancement pattern: Uniformizing Dural tail sign: absent Intratumoral calcification: absent Edge of tumor: multilobular Adjacent bone: eroded	T1W1: Isointensity T2W1: Isointensity Enhancement pattern: Uniformizing Dural tail sign: present Intratumoral calcification: present Edge of tumor: well defined Adjacent bone: thickened

for the differentiation from meningiomas [6]. Interestingly, recent studies revealed that these lesions share a common genetic, pathological, and immunohistochemistry alteration with solitary fibrous tumors of the dura, involving the neuraxis: 12q13 inversions and amplification of the NAB2-STAT6 reporter gene [2]. The World Health Organization (WHO) integrated the 2 terms in 2016 into a single medical condition known as solitary fibrous tumor/hemangiopericytoma (SFT/HPC) and categorized them into 3 grades (Table 1) [3].

M. Fargen et al. reviewed the cases between 1996 and 2010, and they found that males and females are equally affected. Moreover, the tumors typically occur throughout the fifth decade [7]. The signs and symptoms are primarily connected to the tumor's location and size; headaches, focal neurological impairments, increased intracranial pressure, and seizures are the most common clinical manifestations [8].

SFT/HPC tumors frequently exhibit imaging characteristics similar to meningiomas. They are also difficult to differentiate histologically and are commonly misdiagnosed preoperatively, even though meningiomas are significantly more common and less likely to recur or spread extracranially [9,10]. Upon literature review, we collected the similarities and differences between the 2 entities and summarized them based on histological and radiological findings (Table 2).

Surgery is the gold standard of therapy. Complete excision of the tumor is preferable, as inadequate resection can result in the tumor reappearing or remaining cells continuing to proliferate [14]. Our patient pursued surgery and radiation therapy as a personal preference, considering her overall performance status. SFT/HPC has a noticeably high metastatic rate, ranging from 23% to 76%. Additionally, its clinical course is exceedingly aggressive, with recurrence rates ranging from 61% to 76% [15].

Conclusion

This case highlights a rare type of intracranial mass that resembles meningioma on imaging studies. It also illustrates that solitary fibrous tumor/hemangiopericytoma should be kept as a differential diagnosis for brain masses, given its aggressive nature, and its potential for metastasis and recurrence. Early detection and resection can ensure a favorable outcome in the proper clinical setting.

Patient consent

Permission was obtained from the patient's representative for the case report and clinical imaging. We certify that we used entirely anonymized images from which the individual cannot be identified, do not contain any identifying marks, and are not accompanied by text that might identify the individual concerned. Additionally, the patient's names, initials, hospital or social security numbers, dates of birth, or other personal or identifying information was not used or mentioned anywhere in the manuscript.

REFERENCES

[1] Park BJ, Kim YI, Hong YK, Jeun SS, Lee KS, Lee YS. Clinical analysis of intracranial hemangiopericytoma. J Korean Neurosurg Soc 2013;54(4):309–16. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3841273/. doi:10.3340/jkns.2013.54.4.309.

[2] Bouvier C, Métellus P, de Paula AM, Vasiljevic A, Jouvet A, Guyotat J, et al. Solitary fibrous tumors and hemangiopericytomas of the meninges: overlapping pathological features and common prognostic factors suggest the same spectrum of tumors. Brain Pathol 2012;22(4):511–21. https://pubmed.ncbi.nlm.nih.gov/22082190/. doi:10.1111/j.1750-3639.2011.00552.x.

[3] Louis DN, Perry A, Reifenberger G, et al. The 2016 World Health Organization classification of tumors of the central nervous system: a summary. Acta Neuropathol 2016;131(6):803–20. https://pubmed.ncbi.nlm.nih.gov/27157931/. doi:10.1007/s00401-016-1545-1.

[4] Davanzo B, Emerson RE, Lisy M, Koniaris LG, Kays JK. Solitary fibrous tumor. Transl Gastroenterol Hepatol 2018;3:94. doi:10.21037/tgh.2018.11.02.
[5] Editor (s) McKeever Paul E. Chapter 20 - Immunohistology of the nervous system. In: Dabbs David J, editor. Diagnostic immunohistochemistry W.B. Saunders; 2010. p. 820–89. ISBN 9781416057666. doi:10.1016/B978-1-4160-5766-6.00024-8.
[6] Carneiro SS, Scheithauer BW, Nascimento AG, Hirose T, Davis DH. Solitary fibrous tumor of the meninges: a lesion distinct from fibrous meningioma. A clinicopathologic and immunohistochemical study. Am J Clin Pathol 1996;106(2):217–24. PMID: 8712177 https://pubmed.ncbi.nlm.nih.gov/8712177/ . doi:10.1093/ajcp/106.2.217.
[7] Fargen KM, Opalach KJ, Wakefield D, Jacob RP, Yachnis AT, Lister JR. The central nervous system solitary fibrous tumor: a review of clinical, imaging and pathologic findings among all reported cases from 1996 to 2010. Clin Neurol Neurosurg 2011;113(9):703–10. Epub 2011 Aug 26. PMID: 21872387 https://pubmed.ncbi.nlm.nih.gov/21872387/ . doi:10.1016/j.clineuro.2011.07.024.
[8] Ma L, Wang L, Fang X, Zhao CH, Sun L. Diagnosis and treatment of solitary fibrous tumor/hemangiopericytoma of central nervous system. Retrospective report of 17 patients and literature review. Neuro Endocrinol Lett 2018;39(2):88–94. https://pubmed.ncbi.nlm.nih.gov/30183202/#:~:text=Clinical%20manifestations%20of%20CNS%20SFT,seizures%2C%20and%20focal%20neurological%20deficits.
[9] Macagno N, Figarella-Branger D, Mokthari K, Metellus P, Jouvet A, Vasiljevic A, et al. Differential diagnosis of meningeal SFT-HPC and meningioma: which immunohistochemical markers should be used? Am J Surg Pathol 2016;40(2):270–8. doi:10.1097/PAS.0000000000000526.
[10] Yamashita D, Suehiro S, Kohno S, et al. Intracranial anaplastic solitary fibrous tumor/hemangiopericytoma: immunohistochemical markers for definitive diagnosis. Neurosurg Rev 2021;44:1591–600. doi:10.1007/s10143-020-01348-6.
[11] Lahlou G, Rodallec M, Nguyen Y, Sterkers O, Kalamarides M. How to radiologically identify a spontaneous regression of sporadic vestibular schwannoma? PLoS One 2019;14:e217752. doi:10.1371/journal.pone.0217752.
[12] Yue X, Huang J, Zhu Y, Du Y. Solitary fibrous tumor/hemangiopericytoma in the cerebellopontine angle mimicking vestibular schwannoma: a case report and literature review. Medicine (Baltimore) 2020;99(13):e19651. doi:10.1097/MD.0000000000019651.
[13] Okubo T, Nagoshi N, Tsuji O, Tachibana A, Kono H, Suzuki S, et al. Imaging characteristics and surgical outcomes in patients with intraspinal solitary fibrous tumor/hemangiopericytoma: a retrospective cohort study. [published online ahead of print, 2021 Mar 11]. Global Spine J 2021. doi:10.1177/2192568221994799.
[14] Champeaux C, Rousseau P, Devaux B, Nataf F, Tauziède-Espariat A. Solitary fibrous tumours and haemangiopericytoma of the meninges. A retrospective study for outcome and prognostic factor assessment. Neurochirurgie 2018;64(1):37–43. doi:10.1016/j.neuchi.2017.10.004.
[15] Kim JH, Jung HW, Kim YS, Kim CJ, Hwang SK, Paek SH, et al. Meningeal hemangiopericytomas: long-term outcome and biological behavior. Surg Neurol 2003;59(1):47–54. doi:10.1016/s0090-3019(02)00917-5.